

Neurodegeneration and Mirror Image Agnosia

Sadanandavalli Retnaswami Chandra, Thomas Gregor Issac¹

Departments of Neurology, and ¹Clinical Neurosciences National Institute of Mental Health and Neurosciences, Bengaluru, Karnataka, India

Abstract

Background: Normal Percept with abnormal meaning (Agnosias) has been described from nineteenth century onwards. Later literature became abundant with information on the spectrum of Prosopagnosias. However, selective difficulty in identifying reflected self images with relatively better cognitive functions leads to problems in differentiating it from non-organic psychosis. **Aim:** In the present study, we investigated patients with dementia who showed difficulty in identifying reflected self images while they were being tested for problems in gnosis with reference to identification of reflected objects, animals, relatives, and themselves and correlate with neuropsychological and radiological parameters. **Patients and Methods:** Five such patients were identified and tested with a 45 cm × 45 cm mirror kept at 30-cm distance straight ahead of them. **Results:** Mirror image agnosia is seen in patients with moderate stage posterior dementias who showed neuropsychological and radiological evidence of right parietal dysfunction. **Conclusion:** Interpretation of reflected self images perception in real time probably involves distinct data-linking circuits in the right parietal lobe, which may get disrupted early in the course of the disease.

Keywords: Data-linking circuits, Degenerative dementia, Mirror image agnosia

Address for correspondence: Dr. Sr Chandra, Department of Neurology, National Institute of Mental Health and Neurosciences, Bengaluru - 560 029, Karnataka, India. E-mail: drchandrasasi@yahoo.com

Introduction

In 1891, Sigmund Freud introduced the term “Agnosia” to explain situations of preserved percept devoid of meaning.^[1] The work of Luria expanded on it, stating the existence of a functional nervous system apart from the structural one.^[2,3] Cognitive distortions, unrealistic reactions, pseudo normalizations, and bizarre perceptions are now known to be organic. We report a group of patients who were treated by psychiatrists for talking to their own image in the mirror, sometimes fighting with those images resulting in injury to self and caregivers as well as damage to the mirror. Faces are biologically very significant and essential for negotiation of the social world. A large number of faces are remembered spatially, temporally, and pictorially, raising a question whether there are discrete circuits for this in the brain.^[1] Reflected self images, photographs, and images of animate and

inanimate objects are distinctly processed. Newborns respond to simple face-like patterns at birth suggesting the presence of face-specific regions at birth itself.^[4] Distinct evoked responses are seen based on the spatial orientation of these patterns.^[5-8] However, a normal human brain does not get confused between real and reflected human images.^[9,10] In 1997, Ramachandran *et al.* reported that in patients with cerebrovascular accidents and visual field neglect, when a mirror was placed in the non-neglected field, they were looking for the objects inside the mirror and called it mirror agnosia.^[11] Ajuriaguerra *et al.* in 1963 described the same in early dementia and mirrored self misidentification delusion in late dementia.^[12] Selective impairment of recognition of faces and visual learning is described in right parietal and bilateral temporal lobe damage.

Non-syndromic hereditary prosopagnosia is also reported.^[13] In this background, we decided to analyze patients who showed strange behaviors with their mirror images.

Patients and Methods

Patients were recruited from the Neurology outpatient department of National Institute of Mental Health and

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Neurosciences, Bangalore, Karnataka, South India. Patients with degenerative dementia with Hindi Mental Status Examination (HMSE) score of >20 and history suggestive of problems in identifying reflected self images, such as unusual attraction, talking, or any kind of interaction with mirror, were evaluated in detail including praxis and gnosis. Thereafter, a simplified face recognition test was used. Postcard-sized black and white pictures of two famous faces (Mahatma Gandhi and Jawaharlal Nehru), two familiar faces (spouse/sibling, child/carer), novel faces (surgeon and terrorist), crying (faces of two crying children), and laughing (faces of two laughing children) were shown to the patient in a comfortable seated position and response elicited. Also, photographs of reflected faces (faces of patient/spouse/carer of at least one year duration) and photographs (patient and close family member) were shown to the patients. In addition, their ability to identify ornaments, objects, and dresses, and their reaction to the image and ability to improve with repetition were tested. Pictures contained only faces and were shown in the sequential pattern. Then they were shown a 45 cm × 45 cm plain mirror kept straight in front of their face at about 30-cm distance. Their ability to recognize the mirror was confirmed by their description of the frame and mirror. Their impression and reaction about the reflected image of themselves and their relatives/carers while in the front of the mirror as well as when away was recorded. Patients whose scores were less than 20 with multiple other neurocognitive defects due to non-degenerative insults and who did not have problems in reflected self image processing were not analyzed. The Hindi Mental Status Examination (HMSE) is the modified version of the Mini Mental Status Examination adapted for illiterate Hindi-speaking populations, which was developed by INDO-US Cross-national Dementia epidemiology study with a total score of 31 and grouped as mild, moderate, and severe with scores 23-26, 19-22, and <18, respectively, (sensitivity 94%, specificity 98%).^[14] The NIMHANS neuropsychological battery was used to categorize the type of dementia. It was considered as

normal or abnormal by comparing with the normative data for age-, gender-, and education-matched control data available^[15,16] [Table 1].

Magnetic resonance imaging (MRI) was done using 1.5 Tesla, T1, T2, and FLAIR image sequences, and this was reported by a qualified radiologist. All patients underwent neuropsychological assessment and all mandatory investigations such as HIV, VDRL, B12, renal, hepatic, and thyroid function, tests for praxis, neglects, and gnosis. All patients were started on disease-modifying drug Donepezil, which was started as 2.5 mg and increased to 10 mg as four of them had Alzheimer's and one had Diffuse Lewy Body disease.^[17]

In total, 69 patients were studied with 65.2% males and 34.8% females and mean age of 62.5 years. Out of these, 24.64% had Fronto Temporal Dementia, 21.84% cases had Vascular Dementia, 20.29% had Alzheimer's, 14.49% were Unclassified and mixed dementia, respectively. About 2.89% had DLBD and 1.45% had Cortico Basal ganglia Degeneration. Mirror agnosia was seen in four patients with Alzheimer's and mirror image agnosia in five patients.

Patient 1

A 62-year-old female with moderate Alzheimer's dementia without any apraxias, aphasias, neglects, or focal neurological deficits. She was normal in the face recognition tasks except with her own reflected image. She did not recognize her mirror image, instead thought of it as a live person who does not speak. Repeatedly, she was asking for the name of the person and finally declared 'here is a woman who does not know her name'. She did not carry any delusional thoughts about the person in the mirror while not in front of the mirror but always wanted to rush to the mirror as if there was a compulsion or pleasure talking to the person in the mirror. She will quickly answer to the people around, had unusual attraction to the mirror, and ignored the physician and people around. She could correctly

Table 1: Neuropsychological tests done

Patient number	Motor speed	Mental speed (Digit symbol substitution test)	Category fluency (animal names)	Working memory (verbal N-Back tests) planning (Tower of London) set shifting (Wisconsin card sorting test) response inhibition (Stroop test)	Verbal comprehension (Token test)	Verbal learning and memory (Auditory verbal learning test)	Visuospatial construction (Complex figure test)
1.	Normal	Impaired	Normal	Normal	Normal	Impaired	Grossly Impaired
2.	Normal	Impaired	Normal	Normal	Normal	Impaired	Grossly Impaired
3.	Normal	Impaired	Normal	Normal	Normal	Impaired	Grossly Impaired
4.	Normal	Impaired	Normal	Normal	Normal	Impaired	Grossly Impaired
5.	Normal	Impaired	Normal	Normal	Normal	Impaired	Grossly Impaired

identify the reflected face of her daughter-in-law, the resident, and her ornaments and dress. She gave no answer with regards to the identity of the person in the mirror. She appeared quite hostile to the woman in the mirror and preferred to go towards the mirror and quarrel with the 'woman in the mirror' demanding to know her name and who she was.

Patient 2

An 80-year-old patient with features of Diffuse Lewy Body dementia. She exhibited only reflected self image recognition problem. She had two different reactions. First, on seeing her image in the mirror she misidentified it to be a Goddess and prayed in the traditional way. Then, on closely looking at the image, she recognized that her ornaments and dress were worn by the image. This led to the second phase when she stopped worshipping and started shouting and fighting asking for her belongings to be returned. She exhibited anger towards her mirror image and accused her of stealing her dress and ornaments. She could not be pacified even with repeated efforts and conveyed to the resident and others around to catch the 'thief' and retrieve her valuables. We attempted fMRI using the photo of the patient as well as that of the reflected image as test paradigm but were not successful in creating artifact-free data.

Patient 3

She was a retired Major from the army and had features of Alzheimer's disease. She was found to be fine with our face recognition tests except for her own image in the mirror. She presumed the image to be alive and interpreted the image as a 'thief'. She was trying to chase the thief by kicking at the mirror. When she was told that it is her own reflected image, she vehemently denied and insisted that the image is a 'thief' and tried to catch hold of the 'thief', but did not express any reaction to the 'thief' in the mirror when moved away from the mirror.

Patient 4

She was a 69-year-old found to be suffering from Alzheimer's disease. Relatives noted she was discussing her family matters with her own image. When asked, she declared the image as an old friend and she had no problems with identification of others in the mirror. While seeing her children directly, she declared that they were trying to cheat her by putting on the appearance of her children but could recognize them when they spoke to her from the next room or when their reflected images were shown. When she was told the images of her children and the people in front belonged to each other, she insisted that 'those people' did not carry the emotions and the mannerisms that were so familiar to her. She vehemently denied that the people in her front

were her own children rather became quite defensive and said that "I am sure that they are not my children at all". (The patient identified the reflected images of her children and by their voice and did not recognize when they came in front of them. The children were seated parallel to the patient in front of the mirror and only the same mirror (45 cm × 45 cm) was used. The patient did feel her old friend next to the image of the children. We did not make her speak to the children in this context when she was asked to identify the reflected image. She felt that the reflected image had all the characteristics of her children. When the children were in the different room, her ability to recognize was inferred by the way she responded to their oral communication from the next room. They were enquiring about general matters such as "How she's doing? What about her food? and their personal matters with reference to routine agricultural activities in the house. To these questions, she relied quite coherently and relevantly without showing any features of disinterest or suspicion but with a sense of authority and experience, which was reconfirmed by her relatives who were quite familiar with her similar premorbid conversational manner. She was started on anti-cholinesterases and after 12 months, the aforementioned symptom did not persist, but there was global deterioration and patient passed away due to coronary artery disease after two and half years.

Patient 5

A 74-year-old farmer from Bangladesh whose illness started as memory disturbances and later developed way finding problems and confusion in dressing. In addition, he had consistent difficulty in identifying his own reflected image and claimed it to be a friend but he did not initiate conversation or fight. He remained impassive and did not initiate any quarrel towards his mirror image. When asked about the image, he replied that 'it was an old neighbor whom he knew quite well and also had seen him several times in the mosque'. He specifically did not make any comment about the dress of the image. He had difficulty once in identifying the reflected image of his son, which improved with repetition.

Results and Discussion

Out of the total 69 patients with dementia studied, five patients had mirror image agnosia for reflected self images. Their average HMSE score was 22.8. Four of them were suffering from Alzheimer's disease and one from diffuse Lewy Body disease. Details are provided in Table 2. It was observed that all these patients had no difficulty in identifying the mirror, identifying reflected images of familiar face except one patient who improved with suggestion. There was no difficulty in identifying inanimate objects and animals or in spatially

Table 2: Clinical and Radiological Details

Gender	Age	Diagnosis	Praxis	Difficulty in recognizing others	Mirror image agnosia for self-image	Reflected image of others	Imaging features	HMSE	Follow up
F	62	AD	Normal	—	+	—	Temperoparietal Right > left	23	Unchanged
F	80	DLBD	Normal	—	+	—	Diffuse	20	Unchanged
F	73	AD	Normal	—	+	—	Right parietal	24	Unchanged
F	69	AD	Normal	+	+	—	Right parietal	23	Improved
				Ability to recognize by visual modality absent but auditory modality present (Capgras Phenomenon present)					
M	74	AD	Affected	—	+	+	Right parietal	20	Unchanged

M = male, F = female, AD = Alzheimer's disease, DLBD = Dementia of Lewy Body Disease, + denotes Presence and – denotes absence

locating images in the mirror. Classical mirror agnosia is described in literature in patients with cerebrovascular accidents having neglects of one-half. They were found to have difficulty in spatially locating reflected images and looked for them inside the mirror whereas they had no difficulty in the reflected images of self, familiar, or inanimate objects but apraxias were more common. In our patients with Degenerative dementia, mirror agnosia was tested in the conventional way, after confirming their visual acuity with appropriate glasses. They were made to identify the mirror. Then, they were given common objects such as comb, pen, mobile phone, camera, and book and asked to identify. Thereafter, they were asked to identify the reflected images of these objects. If everything was done correctly, the patients were asked to locate the objects tested by looking at the reflected images. Instead of looking for them in the right place, these patients plunged their hands into the mirror, tried to lift the mirror, or went behind the mirror looking for these objects. The group who had mirror image agnosia did not have any feature of the above mirror agnosia. One had apraxia and all showed the common feature of misidentifying reflected self images resulting in varying kinds of misinterpretations as friend, God, thief, or stranger, and the subsequent interaction was based on the faulty perception.^[18] Their everyday life became affected as they showed a pathological attraction to the mirrors and broke mirrors resulting in injury to themselves and damage to the mirrors. Their personal life was affected as they were suspected to suffer from psychiatric illness, reacting and conversing with the image in the mirror. All of them were treated with antipsychotics without any beneficial effect but had side effects. Their families were shattered by the understanding that the head of their family had developed a psychiatric illness. On the other hand, if the organic nature had been identified, it could have been used as a phenotypic marker to decide appropriate tests to diagnose the underlying

organic problem and initiate early treatment. All these patients were in moderate dementia, and they were not dependent for their activities of daily living. One patient improved with medication though other parameters deteriorated. We feel that reflected self images are unique because the individual has seen his own face as a reflected image only and stored as body schema in the non-dominant parietal lobe, whereas the faces of others are both seen as the real as well as in the virtual form and during retrieval believed to be matched with pre-existing schema. Previous studies suggest that there is highly specific representations for faces showing various emotions, primate versus man, inverted versus upright, and living versus non-living. Therefore, it is likely that reflected self images too have specific representations probably in the non-dominant parietal lobe. This is also explained by the typical absence of the classical mirror agnosia and other apraxias in this group of patients at that point of time when they showed predominantly mirror image agnosia for reflected self images, resulting in them being labeled as suffering from psychotic illness until they were submitted for detailed neuropsychological assessment and the nature of the disease was confirmed. As the disease advanced, the above symptom improved in one patient, whether it is due to selective recovery due to the pharmacotherapy or global deterioration masking the phenomenology is not clear. As previously reported by Nora Breen and colleagues (2001), our patients could also identify themselves and others in photographs, and they postulated that reflected image recognition involves representation of oneself in the present time and may be impaired selectively in the presence of face processing impairment, and probably self recognition in mirror and photographs are dissociable.^[19] A similar phenomenon is also reported as “mirrored-self misidentification delusion” (historically, it has also been referred to as the “mirror sign” as well). Mirrored-self misidentification is the belief that one's reflection in the mirror is not

oneself. It has previously been reported many times and in many different clinical conditions, especially following cerebrovascular events and neurodegenerative diseases.^[12] Our patients are unique in the sense that they lacked persistent delusional thoughts about the reflected images of self, had no problem in test of face recognition, and suffered from degenerative dementia and not vascular. They behaved as if the reflected image was real, and one patient had a compulsive pleasurable attraction to talk to the reflected image. We feel this condition involves impairment in reflected self image processing and probably not a delusion. Reflected self images are special as no individual has ever seen his or her real image unlike the images of others, and self images are seen only as reflected images. Neuroimaging showed right parietal atrophy in three patients and diffuse atrophy including right parietal atrophy in two patients [Figure 1]. Further evaluation of a larger number of patients with PET and FMRI is likely to yield insights into the manner in which the brain processes mirror images. A high degree of suspicion and proper assessment might help physicians to recognize the organic cause of the symptom so that early therapeutic interventions can be initiated. The attributes added to the reflected self image as 'friend', 'thief', 'God', or just a 'Stranger' seems to be related to their premorbid personality and occupation such as the army personnel misidentifying as 'thief' and the housewife as 'God'.

Conclusion

The observations of problems in patients in processing reflected images suggest that reflected self images are processed in a different way, unlike real images. This could be altered by parietal lobe disease. People are believed to use different modules for spatial orientation and recognition of reflected self images and probably the co-ordinate system is different for real time processing, which happens in reflected self

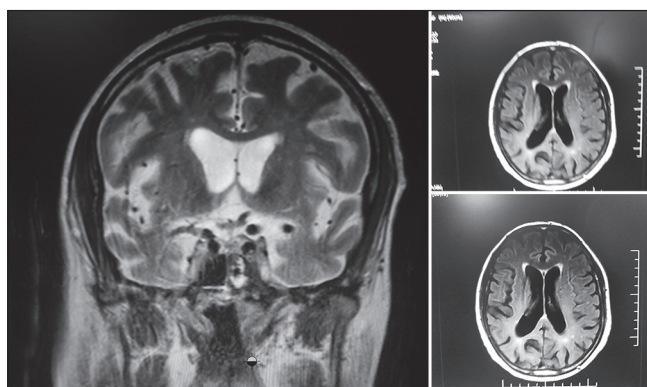


Figure 1: T1 and T2 weighted images show right lateral ventricle larger than left with diffuse atrophy. Insula and parietal regions show more widening of sulci on the right compared to left

images as against photographs. Literature has evidence for defects in spatial orientation of reflected images in patients with neglects due to focal brain insult due to cerebrovascular accidents and neurodegenerative diseases such as dementias resulting in defective mirrored self identification. Connors *et al.* (2014) also managed to recreate mirrored self misidentification delusion in healthy participants by disrupting face processing or mirror knowledge as well as belief evaluation, using hypnotic suggestion.^[20] Our patients showed defective reflected self image processing in the absence of focal brain insults in the form of neglects or any evidence of other forms of face processing defects except in one patient. Neuropsychological tests as well as neuroimaging showed consistent abnormality in the non-dominant parietal lobe. This probably indicates that there are specific circuits for real-time reflected image processing, and they can be selectively affected in the course of neurodegeneration. The content of their delusion seems to be reflected based on their pre-morbid education and occupation.

References

1. Kirshner HS. Approach to Common Neurological Problems. Part 1, Ch 11. The Agnosias. p. 133-9.
2. Ramachandran VS. Illusions of body image: What they reveal about human nature. In: Linas R, Churchland P, editors. The Mind-Brain Continuum. Cambridge: MIT Press; 1996. p. 29-60.
3. Riddoch G. Visual disorientation in homonymous hemifields. Brain 1935;58:376-82.
4. de Haan M, Pascalis O, Johnson MH. Specialization of neural mechanisms underlying face recognition in human infants. J Cogn Neurosci 2002;14:199-209.
5. Aylward EH, Park JE, Field KM, Parsons AC, Richards TL, Cramer SC, *et al.* Brain activation during face perception: Evidence of a developmental change. J Cogn Neurosci 2005;17:308-19.
6. Eimer M, McCarthy RA. Prosopagnosia and structural encoding of faces: Evidence from event-related potentials. Neuroreport 1999;10:255-9.
7. Tulving E, Kapur S, Craik FI, Moscovitch M, Houle S. Hemispheric encoding/retrieval asymmetry in episodic memory: Positron emission tomography findings. Proc Natl Acad Sci USA 1994;91:2016-20.
8. Eimer M. Effects of face inversion on the structural encoding and recognition of faces. Evidence from event-related brain potentials. Brain Res Cogn Brain Res 2000;10:145-58.
9. Haxby JV, Ungerleider LG, Horwitz B, Maisog JM, Rapoport SI, Grady CL. Face encoding and recognition in the human brain. Proc Natl Acad Sci USA 1996;93:922-7.
10. Carroll L. Through the looking glass. In: The complete works of Lewis Carroll. New York: Barnes & Noble; 1994.
11. Ramachandran VS, Altschuler EL, Hillyer S. Mirror agnosia. Proc Biol Sci 1997;264:645-7.
12. Connors MH, Coltheart M. On the behaviour of senile dementia patients vis-à-vis the mirror: Ajuriaguerra, Strejilevitch and Tissot (1963). Neuropsychologia 2011;49:1679-92.

13. Kennerknecht I, Grueter T, Welling B, Wentzek S, Horst J, Edwards S, *et al.* First report of prevalence of non-syndromic hereditary prosopagnosia (HPA). *Am J Med Genet A* 2006;140:1617-22.
14. Tiwari SC, Tripathi RK, Kumar A. Applicability of the Mini-mental State Examination (MMSE) and the Hindi Mental State Examination (HMSE) to the urban elderly in India: A pilot study. *Int Psychogeriatr* 2009;21:123-8.
15. Rao SL, Subbakrishnan DK, Gopulkumar K. NIMHANS Neuropsychological Battery. Bangalore: NIMHANS Publications; 2004.
16. Tripathi R, Kumar JK, Bharath S, Marimuthu P, Varghese M. Clinical validity of NIMHANS Neuropsychological Battery for Elderly: A preliminary report. *Indian J Psychiatry* 2013;55:279-82.
17. Birks J. Cholinesterase inhibitors for Alzheimer's disease. *Cochrane Database Syst Rev* 2006;CD005593.
18. Chandra SR, Issac TG. Mirror image agnosia. *Indian J Psychol Med* 2014;36:340-3.
19. Breen N, Caine D, Coltheart M. Mirrored-self misidentification: Two cases of focal onset dementia. *Neurocase* 2001;7:239-54.
20. Connors MH, Barnier AJ, Coltheart M, Langdon R, Cox RE, Rivolta D, *et al.* Using hypnosis to disrupt face processing: Mirrored-self misidentification delusion and different visual media. *Front Hum Neurosci* 2014;8:361.

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